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Discussion

Dr Marshall L. Jacobs *(Philadelphia, Pa).* Thank you, Dr Pigula, for sharing this wealth of information with us. To you and your associates, congratulations on the very fine results with these difficult patients.

In 2002 I had the privilege of discussing a paper from the same institution titled ''Improving Results of the Modified Fontan Operation in Patients with Heterotaxy Syndrome'' (Ann Thorac Surg. 2002;74:1967-78). Dr Christof Stamm at that time reported the results of 135 Fontan procedures. Nearly three fourths of them were in the decade between 1990 and 2000. Right isomerism predominated over left, which was the opposite of your pattern of predominance in the current series. For patients after 1991, in the Fontan series, the early mortality was just 3%, which is very similar to yours, and the Kaplan–Meier 10-year survival was 93%, which is identical to what you have shown. The risk factors for mortality were anomalous pulmonary venous connection and increased pulmonary vascular resistance. Not surprisingly, in the Fontan group, the previous bidirectional Glenn anastomosis had a positive impact on survival.

Today you have shown us the other side of the coin—91 heterotaxy patients who underwent two-ventricle repair during a time interval when, according to your manuscript, an additional 280 heterotaxy patients underwent Fontan operations. The only risk factor in your series for mortality was unbalanced AV canal defect. The survivors tended to be younger patients. Freedom from arrhythmia was 60% at 5 years and 55% at 10 years. Freedom from reoperation and reintervention were actually considerably less for the biventricular repair group than for the Fontan cohort if one excludes device closure of fenestrations.

At follow-up in your series, the vast majority of patients undergoing biventricular repair were in NYHA functional class I, despite the relatively high incidence of reinterventions and arrhythmias. This was considerably higher than the fraction of the Fontan patients in NYHA class I.

My first question is as follows:

Your manuscript includes the statement: ''Patients with unbalanced complete atrioventricular canal are a high-risk group for which selection criteria are particularly important.'' You point out that the patients in that diagnostic group were believed by the cardiologists to have mildly unbalanced CAVC, and only 1 patient had undergone a bidirectional Glenn before biventricular repair. Another patient underwent a bidirectional Glenn as part of a subsequent re-repair or bailout. What criteria would you now apply to determine the best pathway for patients with mild ventricular imbalance? If those criteria are still illusive, might there be a greater role for staging with a bidirectional Glenn that could culminate in a biventricular repair or even a one-and-a-half ventricle repair?

Dr Pigula. Thank you for your comments. You are absolutely right. The unbalanced ventricle has been the problematic patient group, and it is not problematic just in this situation, of course.

When we evaluate these patients, we look at the typical anatomy, or the typical components of the anatomy. For biventricular repair we should require that the ventricle be of adequate size, possess adequate function, have adequate dedicated AV valve tissue, and adequate outflow tracts.

Now, quantifying those points is where it becomes difficult. That is where an overall evaluation of the composite anatomy, rather than the component anatomy, gets to be difficult. It is a matter of judgment in most cases. The fundamental issue with this kind of study, as well as the previous Fontan study out of our institution with heterotaxy, is patient selection. That is something that this paper did not address. It is a very difficult topic to try to quantify and report on, and that is why we have not been able to do that. However, I think the fundamental issue is which patients are directed toward which treatment strategy. I think that that is really exposed with the unbalanced canal defect because that is where the patients are at risk. Thus I do not have a discrete or a definite answer for you in terms of using criteria A, B, and C, but rather a judgment about the aggregate anatomy.

Dr Jacobs. I have just one other question. In your arrhythmia follow-up data in the paper, you mentioned that there were a number of patients who had either ventricular tachycardia or ventricular fibrillation and required radiofrequency ablation or AICD. Were those late postoperative events? Is that an ongoing risk factor for these patients? Is there any evidence that it is related to septal resection or any other aspect of septation for biventricular repair?

Dr Pigula. I do not know the answer to that, but my instinct/intuition about it is that it is related to the amount of intracardiac work. If you compare the two series, the Fontan series and this series, certainly our freedom from arrhythmia is less than we see in the Fontan group. I think that is probably related to the amount of intracardiac work, in some cases VSD enlargement or incisions in the ventricle, and I think all of those add to it. I do think that they are going to have a greater degree of electrical instability over the long term and that will continue.

Dr Hiromi Kurosawa *(Tokyo, Japan).* Dr Pigula, right isomerism heart, so-called asplenia, usually has a dominant right ventricle

with common AV valve. According to your abstract, 9 patients had right isomerism. Did you do the biventricular repair for those patients who had asplenia with common AV valve?

Dr Pigula. Most of those patients had a common AV valve. I think it was 6 patients who had a common AV valve. Also, several of those patients had L-transposition of the great vessels as well.

Dr Kurosawa. Usually the asplenia heart has a very small left ventricle and a large right ventricle, so that it is sometimes difficult to divide the right ventricle into 2 ventricles.

Dr Pigula. I think your question is septating the AV valve in this setting?

Dr Kurosawa. Yes, and also the ventricle.

Dr Pigula. Well, of course, it gets back, again, to the issue of patient selection. These patients were only accepted for biventricular repair if we thought they had septatable ventricles that we could connect to their appropriate outflow tract and a septatable valve.

I think an important point is that you can have components, those components, the AV valve, the ventricular mass and size, and the outflow tracts, that are all adequate in isolation, but when you put together a composite anatomy, they still may not add up to a biventricular repair. You may still have cases in which you have functional components, but in aggregate they are not amenable to biventricular repair. These were carefully selected patients in whom we thought we could achieve a biventricular repair.